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Case Report

Trabeculectomy with ExPress – an Effective Solution to Posner-Schlossman Syndrome

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Abstract

Posner-Schlossman syndrome (PSS) is a unilateral ocular disease, characterized by recurrent non-granulomatous anterior uveitis and increased intraocular pressure (IOP), leading to chronic secondary glaucoma.

In this case report, we present the case of a 46-year-old man with chronic, hypertensive anterior uveitis in the left eye and frequent recurrences over the last two years. The patient suffers from Posner-Schlossman syndrome with chronic secondary glaucoma, unresponsive to medication therapy. After trabeculectomy with ExPress implantation and, later on, cataract surgery with implantation of an intraocular lens in the left eye, the IOP varied between 10 and 12 mmHg over a period longer than a year without any medication therapy.

Patients with PSS are at risk of developing secondary glaucoma and irreversible complications. Trabeculectomy with ExPress implantation is a reliable and effective treatment of IOP unresponsive to medication therapy.

Keywords

anterior uveitis, Posner-Schlossman syndrome, secondary glaucoma, trabeculectomy

INTRODUCTION

Posner-Schlossman syndrome (PSS) was first described in 1948.[1] It is a rare eye disease, typically observed in young men aged 20-50. It manifests itself as unilateral, non-granulomatous uveitis with increased intraocular pressure (IOP). The disease is thought to be self-limiting, but frequent and recurrent episodes of uncontrollably elevated IOP can lead to the development of open-angle glaucoma.[2,3]

During an attack, patients complain of blurred vision and mild inflammation in the anterior chamber, with the presence of small to medium-sized fine corneal precipitates. The attacks subside spontaneously after a few days to a week and the IOP returns to normal in the periods of remission.[4]

The etiology of the disease is still not fully clarified and studied. Viral infection is supposed to play a significant role. According to various studies, the incidence of the cytomegalovirus (CMV) causative agent isolated from an anterior chamber puncture in patients with PSS ranges from 26% to 62%.[2,5-7] Herpes simplex virus (HSV) and varicella zoster virus (VZV) are less commonly reported as causative agents.[8] Genetic predisposition and gender predilection have been proved, and the prevalence amongst young men is probably determined by the patient's endocrine, humoral, and immune status. Attacks become more frequent in spring. This is the season when asthma and other allergic diseases worsen. There are reports in literature which establish a link between allergic diseases and the development of PSS.[9]

The purpose of this study was to present our therapeutic approach to a patient with PSS.

CASE REPORT

The clinical case we present is that of a 48-year-old man admitted to the University Eye Clinic in Plovdiv, Bulgaria, in September 2017 with an attack of hypertensive, chronic, unilateral iridocyclitis of the left eye. He had first suffered from hypertensive iridocyclitis in his left eye in November 2010. Two years earlier, the same left eye had sustained a non-perforating injury. Between 2017 and 2018, the patient had four consecutive recurrences of iridocyclitis in the left eye. The visual acuity of the left eye for this period (2017-2018), varied from 0.09 to 0.9, and the IOP was within the range of 25–52 mmHg by Goldmann tonometry. The objective finding in the anterior segment of the eye showed: conjunctiva with injection; cornea - single, fine, grayish precipitates, located centrally and in the lower half, with a linear macula below the pupillary plane as a result of a previous trauma; anterior chamber - at a normal depth, showing mild inflammatory cell infiltration, Tyndall of ½ - 1 (+); atrophy of the iris stroma; an open iridocorneal angle was visualized on gonioscopy; a pupil with an oblique oval (due to the trauma), without any posterior synechiae; no pathological changes were found in the fundus. The visual acuity of the right eye was 1.0, with normal IOP values ranging from 10 to 18 mmHg. The anterior segment of the eye and the fundus were free of pathology.

The patient underwent the following laboratory and diagnostic tests: complete blood count, blood sugar level, lipid and kidney profile, ESR, C-reactive protein, antinuclear antibodies, urine test, X-ray of the lungs showing no abnormalities. In order to clarify the etiology, serological tests were performed for toxoplasmosis, tuberculosis, syphilis, HSV, HZV, CMV, and HLA-B27 typing. Of all the tests performed (blood serum), only the values for CMV (IgG) - 290 IU/ml, HSV (IgG) - 8800 IU/l, and VZV (IgG) - 2600 IU/l showed deviation from the reference limits.

Analyzing the data from typical clinical findings, along with the clinical and laboratory results, we established a diagnosis of PSS.

During uveitis attacks, the patient was treated topically with corticosteroids, nonsteroidal anti-inflammatory and antiglaucoma medications. Considering the increased serum titers of the viral agents, treatment with acyclovir was administered: a starting therapy of 2 g/24 h, with gradual dose reduction over a period of four months.

Upon subsequent hospitalization in January 2019, the clinical examination showed secondary glaucoma of the affected left eye (35–55 mmHg according to Goldmann), unresponsive to medication therapy – topically with a beta-blocker, carbonic anhydrase inhibitor, and alpha-2 adrenergic agonist, and systemically with carbonic anhydrase inhibitor 500 mg/24h. Biomicroscopy visualized congestive conjunctival infection, an old corneal cicatrix, iris stroma atrophy, and initial cataract with no evidence of uveitis activity. In the fundus, asymmetry between the two papillae and some thinning of the neuroretinal rim in the left eye were observed ***(Fig. 1)****.*

Specular microscopy examination of the endothelial cells of the cornea showed that in the left eye they had a reduced number, increased size and decreased density in the center of the cornea ***(Fig. 2)****. In the fundus of the left eye, affected by the process, changes in the optic disc were observed – vertical drainage of the excavation* ***(Fig. 1)****. The changes in the left eye optic disc were also confirmed by optical coherence tomography (OCT), which demonstrated sectoral thinning of the ganglion-cell complex and the layer of nerve fibers* ***(Fig. 3)****. A perimeter was marked, showing a paracentral defect in the visual field* ***(Fig. 4)****. Due to the data from the clinical findings and the IOP unresponsive to medication therapy, a decision was made to administer surgical treatment. In February 2019, trabeculectomy with ExPress implantation was performed. It was performed using a standard technique with subconjunctival administration of mitomycin C.*

More than a year later (May 2020), due to clouding of the left eye lens and deterioration of visual acuity, cataract surgery was performed with implantation of an intraocular lens.

After this period, the patient has been monitored every four months so far. The visual acuity of the affected left eye is high (0.9/1.0), the intraocular pressure is maintained within

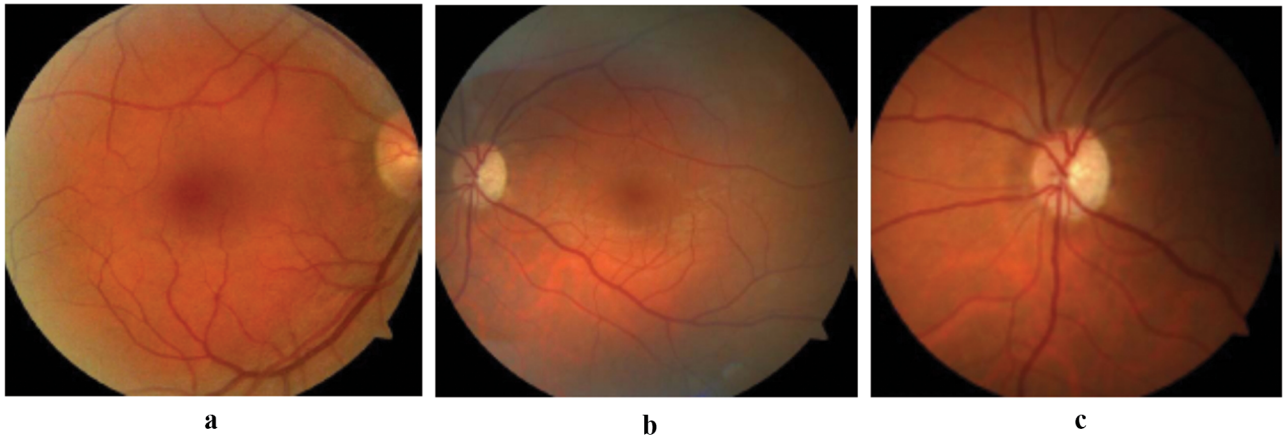
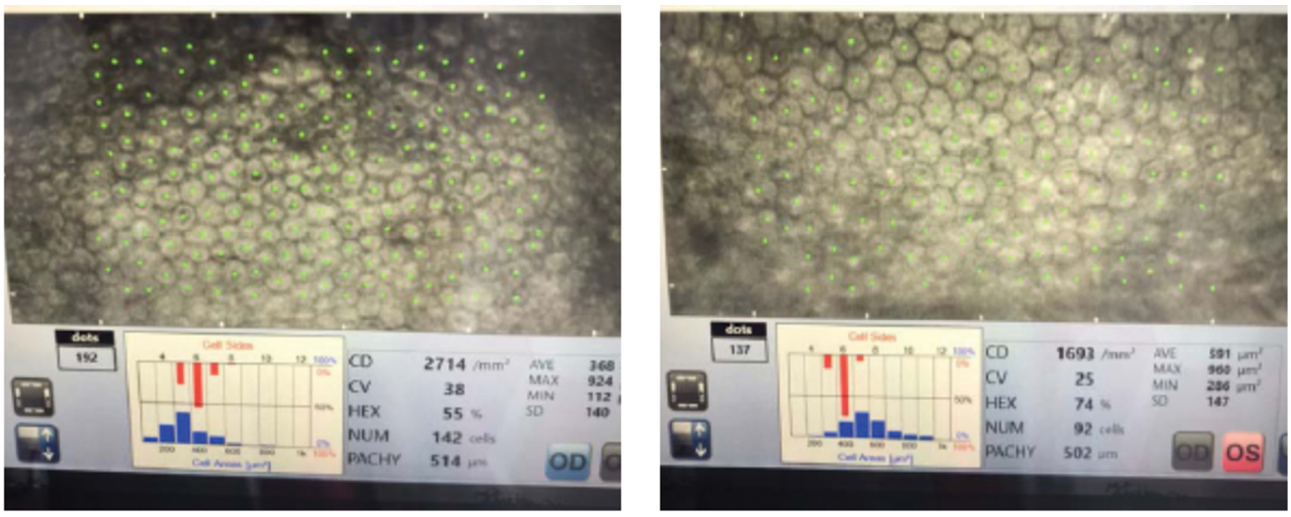
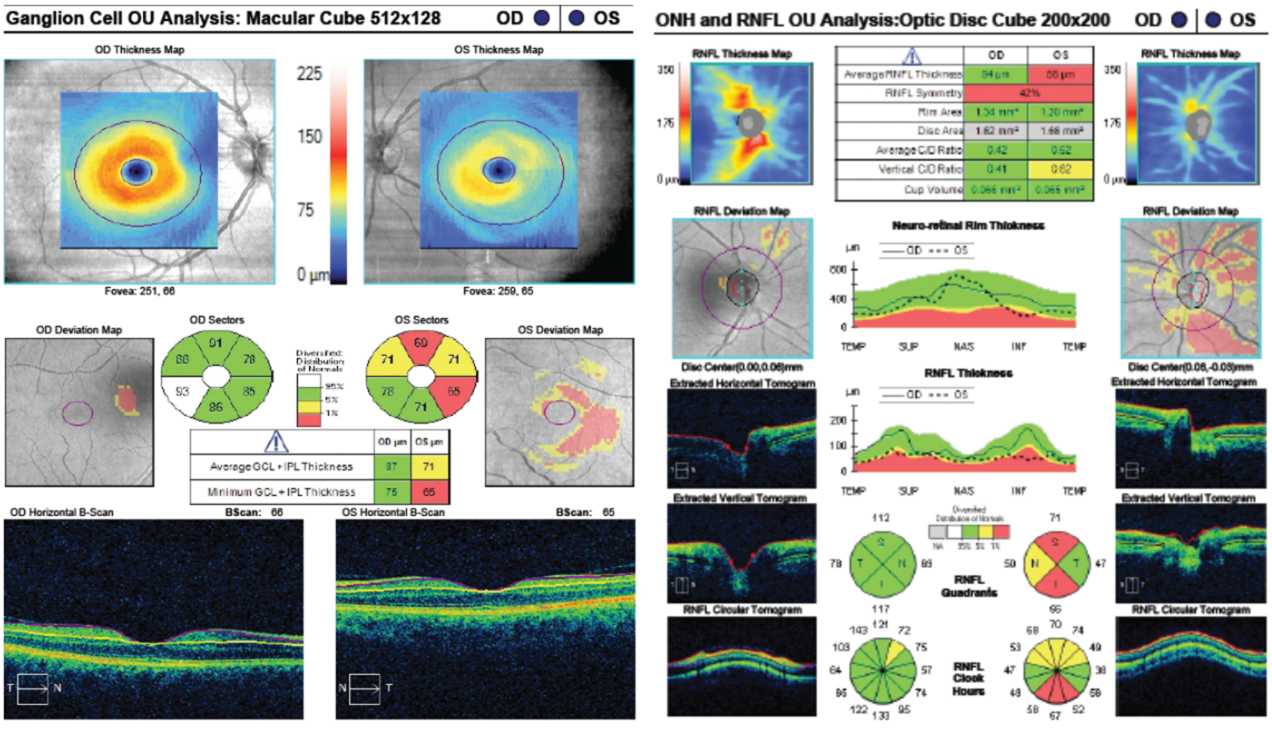


Figure 1. Bilateral fundus photographs (**a, b**) show asymmetry between the two optic disks and thinning of the neuroretinal rim in the left eye (**c**).



a b

Figure 2. Bilateral endothelial cells count (**a, b**) show reduced number, increased size and decreased endothelial cells density in the center of the left eye cornea (**b**).



a b

Figure 3. OCT examination of bilateral ganglion cell complex (**a**) and retinal nerve fiber layer thickness (**b**) shows sectoral thinning of the ganglion cell complex and the layer of nerve fibers in left eye.

normal limits (10-12 mmHg), without the use of antiglaucoma medications and without uveitis activity ***(Fig. 5)****.*

DISCUSSION

PSS is a disease that usually has a favorable course, rarely leading to secondary glaucoma unaffected by medication treatment, or optic glaucoma neuropathy.[10,11] Both the etiology and pathogenesis of the disease are not yet fully clarified. Reactivation of a latent viral infection, leading to an immune response, is thought to play a significant role in the pathogenesis of the syndrome. Viruses have an affinity for centrally located corneal and trabecular endothelial cells, resulting in a decrease in the number of endothelial cells in the center of the cornea and the development of

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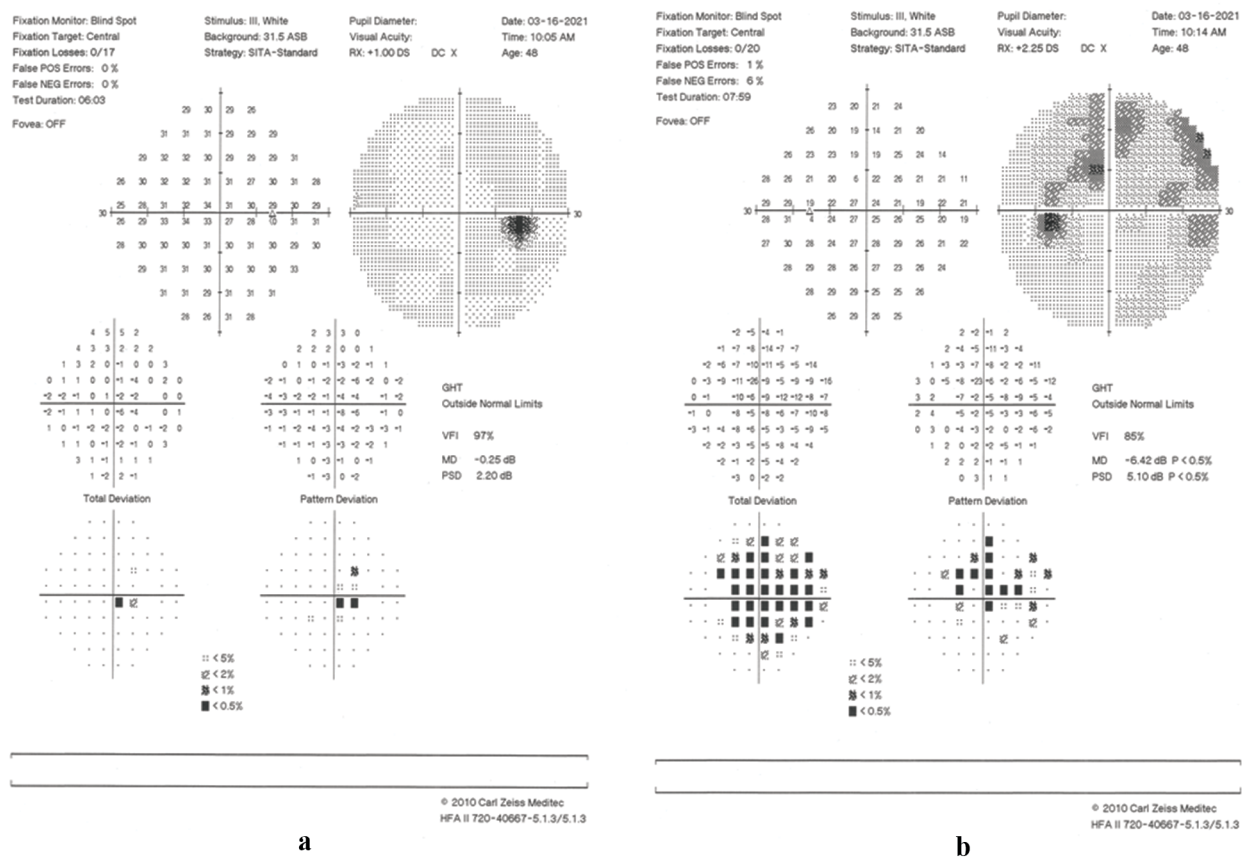

Figure 4. Examination of the 30° visual field of both eyes (**a, b**). A paracentral defect in the upper half of the visual field is visualized in the left eye (**b**).

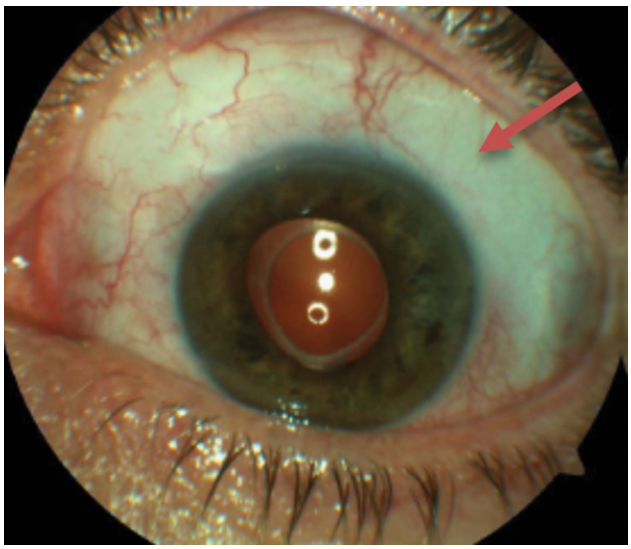

Figure 5. Photograph shows no inflammation of the left eye one year after cataract surgery and more than two years after trabeculectomy. The arrow shows an ExPress implant.

acute trabeculitis at the same time.[2,12] In our clinical case, we also established a reduced number, increased size, and decreased density of the endothelial cells in the center of the cornea of the eye involved in the process. This fact is confirmed by other studies in literature. Setälä and Vanas examined 21 patients with PSS, providing photodocumentation by means of a specular microscope, comparing the density of the centrally located endothelial cells of the affected and the healthy eye.[13] Sixteen of the cases demonstrated repeatedly experienced attacks, the density of endothelial cells being lower in the affected eye. Four patients, after the first attack, and one patient after the second attack did not show any difference in endothelial density of the two eyes. These data confirm that in addition to viral replication, an increase in the number of attacks also increases the risk of developing secondary glaucoma and eye damage. As regards our patient, over a period of about a year, the number of uveitis recurrences has been significant (four). Another study, by Maruyama et al., examined 33 eyes (33 patients) with PSS, 21 of whom underwent surgical treatment. The researchers divided the patients into two groups: the patients in the first group had received only medication therapy and those in the other group had undergone surgical treatment. The authors detected a significant correlation, concerning the reduction in density of centrally located endothelial cells, between the surgically treated group and the medication treated one ($p<0.05$), as well as a significant difference in maximum IOP and the presence of glaucomatous defects in the visual field between the two groups ($p<0.01$). However, they did not discover any cor-

relation between the frequency of attacks and the reduction in the density of endothelial cells.[1]

The serological results showed high levels of IgG for CMV, HSV, and VZV in our patient's blood plasma, due to which we performed antiviral treatment for a period of four months. The majority of studies recommend a three-month general course of antiviral therapy when there is a proven viral agent.[10,14,15] This leads to a significant reduction in viral replication, but not to its complete elimination from the body.[14-16] Some authors recommend that 2% solution of ganciclovir be applied topically in case of a proven CMV infection, which they believe leads to faster control of the uveitis attacks, reducing the risk of damage to corneal endothelial cells.[3]

The inflammatory edema of the trabecular meshwork also plays a key role in the increase in IOP during an acute attack.[16] Yan et al. observed some significant thickening of the trabecular apparatus in PSS eyes compared to contralateral healthy controls.[12] What is also considered to be causing an elevated IOP is the increased level of transforming growth factor β, isolated from the anterior chamber of glaucoma patients. Transforming growth factor β not only increases the amount of extracellular matrix, but also suppresses its disintegration. In addition, the factor increases the contractility of trabeculum cells while, on the other hand, inhibiting cell proliferation and ultimately reducing the number of cells in the trabeculum, which worsens the prognosis for the patient. [2]

According to literature data, there is a 2.8-fold higher risk of developing glaucoma if uveitis persists for over ten years.[2] In the clinical case currently presented, uveitis has lasted for nine years. In patients with PSS, IOP should be actively monitored during uveitis attacks as well as during periods of remission in order to avoid optic nerve damage.[12]

It is not only the duration of the disease that is of importance but also the number of relapses. With our patient, their increased number led to an uncontrollable increase in IOP. The risk factors which predict the need for surgical treatment in patients with PSS are as follows: persistently elevated IOP unresponsive to therapy; the presence of CMV-DNA in puncture at the anterior chamber, as well as frequent recurrences of uveitis.[1] In the case at hand, almost all of the described criteria are present – elevated IOP, changes in the optic disc, combined with sectoral thinning of the ganglion cell complex and the layer of nerve fibers of the retina, as well as changes in the visual field. The appearance of structural changes can lead to a permanent decrease in visual acuity and deterioration of the quality of life of patients at a relatively young age.

An important goal in the treatment of the disease is to bring the elevated IOP under control and to prevent loss of vision.[1] Secondary glaucoma, unresponsive to medication therapy in our case, led to a decision for surgical treatment. Glaucoma surgery is to be preferred in the presence of a high risk of irreversible visual impairment. Trabeculectomy reduces the severity and recurrence of uveitis by draining inflammatory cells from the anterior chamber into the subconjunctival space.[2] We decided to perform trabeculectomy using MMC and antiglaucoma implantation due to the relatively young age of the patient. The increased number of fibroblasts, lymphocytes, and macrophages in the conjunctiva of the eye with uveitis poses a higher risk of postoperative fibrous proliferations.[16] Our decision is shared by other authors too. According to Maruyama et al., patients who do not respond to antiglaucoma therapy and such with progressive glaucoma defects must undergo surgical treatment.[1]

CONCLUSIONS

Patients with PSS and permanently elevated IOP are at high risk of developing secondary glaucoma and irreversible damage. The duration of uveitis, the number of recurrences and the presence of structural changes are essential prognostic factors for the need of surgical therapy. We consider that trabeculectomy with ExPress implantation is a reliable and effective treatment of IOP uncompensated under medication therapy.

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Трабекулэктомия с помощью ExPress – эффективное решение синдрома Posner-Schlossman

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Резюме

Синдром Posner-Schlossman (PSS) представляет собой одностороннее заболевание глаз, характеризующееся рецидивирующим негранулематозным передним увеитом и повышением внутриглазного давления (ВГД), приводящее к хронической вторичной глаукоме.

В этом клиническом случае мы представляем случай 46-летнего мужчины с хроническим гипертоническим передним увеитом левого глаза и частыми рецидивами в течение последних двух лет. Больной страдает синдромом Posner-Schlossman с хронической вторичной глаукомой, не поддающейся медикаментозной терапии. После трабекулэктомии с имплантацией ExPress, а затем операции по удалению катаракты с имплантацией интраокулярной линзы в левый глаз ВГД колебалось от 10 до 12 mmHg в течение более года без какой-либо медикаментозной терапии.

Пациенты с PSS подвержены риску развития вторичной глаукомы и необратимых осложнений. Трабекулэктомия с имплантацией ExPress является надёжным и эффективным методом лечения ВГД, не поддающегося медикаментозной терапии.

Ключевые слова

передний увеит, синдром Posner-Schlossman, вторичная глаукома, трабекулэктомия